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Lymphangiomas are benign, tumors infrequently found in the abdomen and rarely in the retroperitoneum (1-4). There are three reports in the English literature in which a retroperitoneal lymphangioma presented as an inguinal herna (4-6). We are presenting a similar case which to the best of our knowledge is the first reported case from India.

Case Report

A two year old male patient was admitted with complaint of bilateral reducible inguinal herna present since birth. During surgery for herna, a multicystic tumor was found protruding through the left inguinal canal behind the herna sac. Repair of bilateral herna was carried out with maximum possible excision of the tumor through the same incision, and a plan to go for definitive surgery after the biopsy report. On the following day, sonographic examination was carried out to look for left over mass. Examination revealed a fluid filled multiloculated mass in left lumbar and iliac region roughly measuring 5 x 9 cm. The features of the fluid were those of simple fluid that is anechoic with enhanced through transmission and with a sharply defined far wall. The left kidney was normal but pushed medially. All other organs were also normal. Based on the sonographic findings a provisional diagnosis of lymphangioma was kept, which was confirmed by histopathological examination. Histopathology showed small and large cavernous spaces lined by endothelial cells filled with lymph and separated by fibrous connective tissue which was infiltrated by focal areas of lymphocytes confirming the diagnosis of cavernous lymphangioma. Due to reluctance of parents, surgery for retroperitoneal mass was delayed but the child was followed-up. After four months, the patient developed an obvious swelling in the left lumbar region extending to the left flank. Repeat ultrasound examination showed the same findings (Fig. 1).

On repeat surgery, a large irregular cystic mass with interspersed solid areas, occupying the left retroperitoneal space was found. The mass was firmly adherent to the posterior parietal peritoneum, extended from the midline to the left flank and from the diaphragm above to the left inguinal
ligament below. The major part of tumor was removed (Fig. 2) and the posterior wall was destroyed by cauterization. The post operative period was uneventful. Histopathology again confirmed the diagnosis of lymphangioma. The child was followed up till he was four years old; there was no recurrence of tumor.

Discussion

Lymphangiomas are benign tumor classified histologically as being simple (capillary), cavernous or cystic, those in retroperitoneal site being almost always of cystic type(8). Cystic lymphangiomas may be unilocular or multilocular and contain serous or chylous fluid. Although the origin of lymphangiomas is not known they are thought to be developmental abnormalities (hamartomas) rather than true neoplasm(9,10). Of the cystic lymphangiomas 75% occur in neck (cystic hygroma), 20%
are found in the axillary region, the rest 5% arise in mediastinum, mesentry, omentum, retroperitoneum, pelvis, groin, spleen or bone(3,4). Cystic lymphangiomas involving the retroperitoneum most commonly present early in life as a large abdominal mass(10).

Sonographic appearance of a septated cystic mass with clear (anechoic) fluid is characteristics(ll-14). If available, CT examination reveals low attenuation value of the fluid suggesting presence of fat containing fluid thus further helping in diagnosis(ll). Retroperitoneal lymphangiomas usually do not calcify, this feature helps in differentiating them from mature teratoma, other possible cause of unilocular or multilocular cystic mass in retroperitoneum(13,14).

The treatment of choice is complete removal. Urgent operation may be necessary in patients with mediastinal lesion causing progressive dyspnea resulting from sudden enlargement of hygroma from hemorrhage or infection. The operation is essentially conservative, since there is no justification for sacrificing any vital structure to achieve complete removal of this benign lesion. However, recurrence may be noted in some such cases at a later stage. Irradiation is of little value because it rarely produces regression and has its inherent drawbacks(3).

REFERENCES